Society News

European Confederation of Neuropathological Societies

Dear Reader,

Below you will find reports on several activities of Euro-CNS. We are pleased to have a new European Fellow of Neuropathology (EFN) who passed the European Examination in Neuropathology that was held on May 15 and 16 in Zürich. The dates of the next Examination are announced as well. On June 10, Euro-CNS had its Executive Committee and Council meetings in London and a brief report follows below. The preparations for the Euro-CNS training course "Basic Neuropathology" (Aachen, September 24 – 27, 2024) are going well and you can find the detailed program at the end of this Newsletter. The educational board of Euro-CNS is announcing an updated course on Developmental Neuropathology, to be held from January 29 – 31, 2025 in Amsterdam.

On the last page you will find the Quiz #24 which you can also make online on the Euro-CNS website and see your score and answers right away.

We trust you will find this information of interest and hope that you will share our announcements with your peers.

With kind regards,
The Euro-CNS News Team

New European Fellow in Neuropathology: Basma AlYamany

Euro-CNS congratulates Basma AlYamany from Riyadh, Saudi Arabia, as she recently passed the European examination in Neuropathology. She is now a registered European Fellow in Neuropathology. This year's European Examination in Neuropathology was held on May 15 and 16 in Zürich, at the "Universitätsspital Zürich" (USZ), Switzerland. The examiners were Professor Herbert Budka, retired Head of Department of the Institute of Neurology at the Medical University of Vienna, and Professor Tibor Hortobágyi from the host institution in Zürich. Initially, there were three candidates. However, due to withdrawals, only one candidate sat the exam: Dr. Basma AlYamany, who is currently Consultant Neuropathologist at King Fahad Medical City, Riyadh, Saudi Arabia. Earlier, she successfully completed a residency training in neuropathology at the London Health Science Centre, London (OT), Canada. Dr. Basma AlYamany passed every component of the examination. The examiners had declared that they were satisfied that Dr. Basma AlYamany is competent to practice independently as diagnostic neuropathologist, and recommended that she should be elected to the European Fellowship in Neuropathology (EFN). The procedure and the report were supported and approved by all members of the Euro-CNS Board of Examiners. An experience report by Dr. AlYamany follows below.

The next European Examination in Neuropathology will held in London, UK, from May 15 – 16, 2025.

Experience report European Examination in Neuropathology – by Dr. Basma AlYamany

I trained in neuropathology in London, ON, Canada, where I had the opportunity to work with exceptional mentors and gain a comprehensive education in the field. This training prepared me well for the challenges and rigors of the European Fellowship of Neuropathology exam, which I took at the Institute of Neuropathology in Zurich. The exam in Zurich was a great experience overall. The location was easy to find, and the Institute's staff made me feel welcome from the moment I arrived. The exam itself was long and thorough, accurately reflecting the day-to-day challenges that a neuropathologist faces. Despite its difficulty, the structure of the exam was comprehensive and fair. The examiners were incredibly welcoming and friendly. It was a privilege to meet such giants in the field, whose expertise and passion for neuropathology were evident throughout the examination process. Their encouraging demeanor helped ease some of the stress associated with such a rigorous exam. Participating in the EFN exam was a rewarding and enlightening experience. It not only tested my knowledge and skills but also reinforced my dedication to the field of neuropathology. I am grateful for the opportunity and proud to have passed this challenging examination.

Submitted by Basma Mahmoud AlYamany, MBBS, EFN, King Fahad Medical City, Riyadh Second Health Cluster, Riyadh, Saudi Arabia

Euro-CNS Council meeting on June 10, 2024, London

All affiliated societies of Euro-CNS were represented at the hybrid Council meeting on the 10th of June. The meeting was hosted at the UCL Institute of Neurology, the National Hospital for Neurology and Neurosurgery at Queen Square, London. The following societies had one or more Councilors participating in the meeting: The Austrian Society of

Neuropathology, the Baltic Association of Neuropathology, the British Neuropathological Society, the Bulgarian Society of Neuropathology, the Dutch Society of Neuropathology, the French Society of Neuropathology, the German Society of Neuropathology (DGNN), the Hellenic Society of Neuropathology, the Hungarian Society of Neuropathology, the Irish Neuropathological Society, the Italian Society of Neuropathology, the Neuropathology Working Group Belgian Society of Pathology, the Polish Society of Neuropathology, the Portuguese Society of Neuropathology, the Romanian Circle of Neuropathology, the Scandinavian Neuropathological Society, the Slovenian Society of Neuropathology, the Spanish Neuropathology Club, the Swiss Society of Neuropathology and the Turkish Neuropathology Working Group.

It was an excellent occasion to welcome new councilors, and to share new developments that were happening in the various countries and regions. The minutes of the previous Council meeting (September 14, 2023) were approved. There are no changes in the board, but the Executive Committee called for nominations for the Vice-Presidency. This position will become available next year, at the time of the European Congress of Neuropathology (June 2025). Only councilors may be nominated for board positions. The Treasurer, Martin Lammens, gave an extensive financial report, including the bookkeeping and an overview of the member dues. The Editor of Clinical Neuropathology, Christian Mawrin, presented details on the editorial activities, submission numbers and other data as provided by the Publisher. Access to the Journal will be for free for Euro-CNS members until at least the end of 2024. On behalf of the Organizing Committee, Wilfred den Dunnen gave an update on the preparations for the European Congress of Neuropathology in Maastricht (June 11 – 14, 2025) and presented the program-at-a-glance as well as the social activities that are being planned. The International Society of Neuropathology will hold the International Congress of Neuropathology from June 6 – 9, 2027 in Edinburgh, Scotland. The Executive Committee called for applications for the 2029 European Congress of Neuropathology. Societies interested to host the congress were asked to contact the Euro-CNS office. Wilfred den Dunnen, as chair of the Educational Board, reported on the recent Euro-CNS Muscle & Nerve Course (March 14 – 16, 2024, Berlin), and on the upcoming courses, being the Basic Course in Neuropathology (September 24 – 27, 2024, Aachen) and the updated course on Developmental Neuropathology in Amsterdam in January 2025. After this, Chairman of the Examination Board, Tibor Hortobagyi, reported on the proceedings of the European Examination in Neuropathology in 2024, and the plans for 2025 (see detailed report above). Secretary-General Zane Jaunmuktane provided information about interaction with the European Society of Pathology for the European Congress of Pathology (ECP) 2024 in Florence, the ECP 2025 in Vienna, and the ECP 2026 in Stockholm. Euro-CNS has been involved with the (joint) organization of neuropathology related sessions at the ESP congresses for many years in a row already. Regina Reimann and Ekkehard Hewer, Councilors of the Swiss Society of Neuropathology, asked all councilors to reflect on and summarize the current situation and perspectives of epigenetic diagnosis of CNS tumors in their own countries and to provide feedback through an online survey. The results of the survey will then be presented at the next Council meeting. The next meeting of the Euro-CNS Council will take place by the end of November 2024 (virtual), and the next physical meeting will be next year at the occasion of the European Congress of Neuropathology in Maastricht, the Netherlands.

Euro-CNS Basic Course in Neuropathology, September 24 – 27, 2024, Aachen

The basic course in neuropathology is meant for those who do not have the benefit of structured neuropathological training for and in their professional neuropathological activities. The course is also intended for medical specialists from general pathology, neurology, or neurosurgery, who would like to get more insight in the field of neuropathology. We refer to the Euro-CNS website for more information including registration details. You will find the tentative program at the end of this News section. At the moment we still have several seats available.

First announcement of the Euro-CNS CME Course on Developmental Neuropathology, January 29 – 31, 2025, Amsterdam

Introduction and description

The newly updated Euro-CNS neuropathology training course "Developmental Neuropathology" will be held from January 29 – 31, at the Amsterdam UMC in the Netherlands. It consists of lectures with an emphasis on teaching. Practical (microscope-based) sessions are part of the course. Participants will have access to a digital platform with case material. This will enable participants to also evaluate the case material after the course. This 3-days course and workshop will provide practicing (neuro)pathologists, clinicians, and researchers with up-to-date reviews of the classification and the diagnostic problems in the pathology of neurodevelopmental diseases. There will be interactive case discussions, and also participants may send in a case for discussion. The course language is English. We recommend bringing a tablet or laptop for digital slide viewing.

Target group

The course is intended for all involved in the pathological diagno-

sis of neurodevelopmental diseases: trainees and specialists in pathology, neuropathology, neurosurgery, neurology, or neuro-oncology.

Topics include

Clinical signs; Neuro imaging; Normal brain development; Genetics of brain development; Defects in brain growth; Cortical malformation; Degenerative diseases; Infections; Epilepsy; Trauma; Vascular malformation.

Organizers

Eleonora Aronica (the Netherlands), Homa Adle-Biassette (France), Wilfred den Dunnen (the Netherlands).

Venue

Amsterdam University Medical Center, location AMC, Meibergdreef 9, Amsterdam, the Netherlands.
Website: www.euro-cns.org

DFP Literaturstudium

Clinical Neuropathology Volume 43: 3, 2024 – educational summary by I. Fischer and J.A. Hainfellner

Daisuke Tahara et al. report their findings of a neuropathological analysis of the inferior olivary nucleus (ION) in an autopsy series of corticobasal degeneration (CBD). A total of 32 consecutive cases of CBD from a single center were included in the study. The inferior olivary nucleus is connected to the contralateral dentate nucleus (DN) and ipsilateral red nucleus via the dentato-rubro-olivary pathway (Guillain-Mollaret triangle). Since the dentate nucleus is known to be affected in CBD, the authors hypothesized that this could lead to hypertrophic olivary degeneration. Furthermore, an association of hypertrophy of the inferior olivary nucleus (HION) with a clinical PSP-like type was deemed likely. For this study, the ION was assessed on Klüver-Barrera stains in a loupe magnification, and enlargement was semi-quantitatively assessed (grade 0 – none, grade 1 -mild or partial enlargement, grade 2 – apparent and entire enlargement). In addition, atrophy and myelin pallor of the hilum and amiculum of the ION, central tegmental tract (CTT), superior cerebellar peduncle, and hilum of the dentate nucleus, neuronal loss and astrogliosis in the ION and DN were assessed in a similar fashion. The severity of Gallyas/tau-positive lesions in the ION and DN was also semi-quantitatively assessed and documented. For each parameter, the authors provide representative histologic images depicting different severity grades. Of the 32 cases included in this study, HION was observed in 14 cases, of which 8 had bilateral symmetrical hypertrophy. All cases with HION also featured myelin pallor within the amiculum and hilum of the ION and CTT, astrogliosis, and swollen neurons in the ION. Gallyas/tau-positive lesions were noted in the ION of all cases with or without HION. In addition, the majority of cases of HION displayed neuronal loss and hilar atrophy/myelin pallor of the contralateral DN. Of note, the ventrolateral part of the ION and caudal part of the DN were predominantly affected. This could be explained by different lesion distribution in the cerebral cortex, which projects to the ventral part of the ION, and the red nucleus, which predominantly projects to the dorsal part. Fibers from the ventral inferior olivary nucleus in turn project to the caudal and ventral DN.

Concerning the correlation of histologic findings with clinical characteristics, there was no association between HION and clinical subtype of CBD. There was, however, a longer disease duration and lower brain weight in cases with HION. The authors hypothesize that the dentate nucleus may be affected in CBD late in the disease, causing transsynaptic degeneration of the ION. Four of 6 cases in this study with laterality of the HION were associated with contralaterally accentuated rigidity. Hypertrophy of the inferior olivary nucleus has not been previously analyzed in CBD in detail – only few reports of such cases exist.

Ozgur Orhan et al. report a case of polymorphous low-grade neuroepithelial tumor of the young (PLNTY) and give a literature review. The case is that of a 32-year-old male patient presenting with seizures for several months. MRI studies revealed a 22-mm right temporal mass lesion with calcification and focal contrast enhancement. A gross total resection of the tumor was performed. After a follow up-period of 1 year, the patient is free of seizures. Histology revealed a cellular tumor composed of oligodendroglioma-like cells with some nuclear pleomorphism and some spindle-shaped tumor cells, as well as extensive calcification. The tumor cells were positive for GFAP, Olig2, CD34, and BRAF. No expression of IDH1 was seen. ATRX was retained. The Ki67-proliferation index was 4 – 5%. A diagnosis of PLNTY was thus rendered. From a morphological perspective, the differential diagnosis to be considered is wide-ranged and includes oligodendroglioma, dysembryoplastic neuroepithelial tumor (DNT), MAPK pathway altered low-grade glioma, gangliolioma, pilocytic astrocytoma, and pleomorphic xanthoastrocytoma. The authors discuss the findings that allow differentiation of these tumors from PLNTY.

In a table, the authors summarize the literature on this entity available so far, including 17 publications describing a total of 66 cases. While PLNTY usually affects children and young adults, occurrence in older adults has also been reported. Seizures are the most common presenting symptom. The clinical behavior is usually benign, with no recurrence after complete resection. On a molecular level, BRAFV600E mutation is most common, followed by FGFR2-fusions, FGF3 mutations, FGFR rearrangements, BRAF fusion, NTRK2 mutations, and EGFR mutations.

In their case report, Neeraj Singla et al. describe an autopsy case of idiopathic hypereosinophilic syndrome (HES) presenting with a stroke.

The patient was a 62-year-old woman with a history of hypertension and generalized itching associated with episodic skin rashes. On neurological examination, she was noted to be drowsy. MRI imaging revealed multiple infarcts involving bilateral frontal, parietal, and occipital lobes and cerebellum. Blood testing showed leukocytosis with eosinophilia. There was no evidence of parasitic infection or myeloid neoplasia. She was accordingly diagnosed with idiopathic hypereosinophilic syndrome. Treatment included ivermectin, methylprednisolone, hydroxyurea, imatinib, as well as cyclophosphamide. Her eosinophil count was not reduced, and she developed aspiration pneumonia and died. On autopsy, the bone marrow was hypercellular with excess eosinophils and eosinophil precursors. The heart showed biventricular hypertrophy, thrombi in the ventricles, fibrin thrombi in intramyocardial vessels, microscopic infarcts, and eosinophilic endomyocarditis. Fibrin thrombi were also found in the gastrointestinal tract. In addition, there was eosinophilic esophagitis, eosinophilic and aspiration pneumonia with bacterial superinfection, and thrombotic microangiopathy of the kidneys.

Examination of the brain revealed no herniation and no venous thromboses. On coronal sections, there were softened areas in both frontal lobes, the right parietal and left occipital lobes, as well as in the cerebellum involving the dentate nuclei and the cerebellar tonsil. Histologically, these represented acute and chronic infarcts in grey and white matter. Additional infarcts were seen in the midbrain and pons.

Hypereosinophilic syndrome is defined as an absolute eosinophil count of > 1,500/µL for 6 months without any known triggers and evidence of organ damage. A classification as "idiopathic" requires the exclusion of causes such as an underlying neoplasm or a parasitic infection. The organ damage occurs directly by infiltrating eosinophils or by release of inflammatory mediators. Most common clinical manifestations include dermatological, pulmonary, gastrointestinal, cardiac, or neurological symptoms. Cases initially presenting with a stroke are rare, however. The precise pathogenesis of stroke in HES has not been fully elucidated: cardiac thromboemboli, thrombogenicity caused by release of eosinophilic basic protein, direct endothelial damage by eosinophils, and increased blood viscosity are possible mechanisms.

Quiz #24 Clinical Neuropathology

Below you will find Clinical Neuropathology Review Quiz #24, carefully compiled by Dr. I. Fischer (Aarau, Switzerland) and Prof J.A. Hainfellner (Wien, Austria). The questions refer to the educational summary (see text above) and the papers of this issue of *Clinical Neuropathology* (Volume 43, No. 3/2024). We recommend making the quiz online, so that you will see your score and the correct answers right away: **https://www.euro-cns.org/journal/journal-quiz/.**

1. Which of the following genes may show a molecular alteration associated with peripheral eosinophilia?

a – PDGFRA
b – PDGFRB
c – FGFR1
d – JAK2
e – All of the above

2. Which of the following histological features is NOT a typical change in idiopathic hypereosinophilic syndrome?

a – Eosinophilic inflammation
b – Vascular thrombi
c – Infarcts
d – Parasitic infestation
e – None of the above

3. Which of the following histological features may be observed in corticobasal degeneration?

a – Astrocytic plaques
b – Neuropil threads
c – Coiled bodies
d – Neuronal loss
e – All of the above

4. Which of the following molecular alterations has NOT been described in polymorphous low-grade neuroepithelial tumor of the young (PLNTY)?

a – FGFR2-INA fusion
b – FGFR-TACC3 fusion
c – IDH1(R132H) mutation
d – BRAF(V600E) mutation
e – FGFR2-KIA1598 fusion

5. Which of the following tumor types may be considered in the differential diagnosis of PLNTY?

a – Ganglioglioma
b – Dysembryoblastic neuroepithelial tumor
c – Oligodendroglioma
d – Diffuse low-grade glioma, MAPK pathway altered
e – All of the above

Quiz submitted by
Ingeborg Fischer,
Switzerland
reviewed by
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Austria

Upcoming events

European Congress of Pathology
September 7 – 11, 2024,
Florence, Italy
Website:
https://www.esp-congress.org/congress/future-congresses.html

European Basic Course in Neuropathology
September 24 – 27, 2024,
Aachen, Germany
Website: https://www.euro-cns.org

The Northern Lights Neuroscience Symposium
September 26 – 27, 2024, Hanasaari, Finland
Website: https://www.helsinki.fi/en/conferences/northern-lights-neuroscience-symposium-2024

Euro-CNS CME course "Developmental Neuropathology", January 29 – 31, 2025, Amsterdam, the Netherlands
Website: https://www.euro-cns.org

European Congress of Neuropathology
June 11 – 14, 2025, Maastricht, the Netherlands
Website: https://www.euro-cns.org

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